313 Can patient-reported health-related quality of life predict survival in cystic fibrosis?

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Objective: This work aims to investigate the role of patient-reported, health-related quality of life (HRQOL) in predicting survival in cystic fibrosis.
Methods: All patients attended the Adult CF Units in Leeds and Manchester in the UK. HRQoL, demographic and clinical variables were assessed during an outpatient visit during 1996–1997. Clinical and demographic variables were: age, gender, FEV_1% predicted, BMI, access device, *B. cepacia*, diabetes, nutritional and transplant status. HRQoL was measured using the Cystic Fibrosis Quality of Life Questionnaire (CFQoL) which comprises 9 domains: physical functioning, social functioning, emotional functioning, treatments, chest symptoms, future concerns, relationships, body image and career issues. Survival analysis was undertaken using Cox Proportional Hazards Regression in Intercooled STATA 9.
Results: At initial assessment there were 223 patients (age range 14 to 52 years; 121 females). On 31st December 2006, 154 were still alive and the death dates of 66 were known (3 were lost to follow-up). Even in the presence of FEV_1% predicted and all the other demographic and clinical variables, the physical functioning domain of the CFQoL was an important predictor of survival. We are currently investigating possible explanations for this.
Conclusion: Patient-reported physical functioning could be an important predictor of survival, and may provide a significant contribution to clinical monitoring.

314 Turkish translation and validation of revised Cystic Fibrosis Questionnaire (CFQ-R) for children

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Developmental course of chronic diseases in children is influenced by physical, psychological and social issues. Measurement of QoL is quantifying the results of the disease and its treatment on the patient's perception of a satisfying life. Cystic fibrosis (CF) is a chronic disease that involves multiple organ systems. Therefore both the disease itself and its treatment may impair quality of life (QoL) in children. On the other hand, there is no QoL questionnaire for children with CF in Turkish. The aim of this study was to translate the Revised Cystic Fibrosis Questionnaire (CFQ-R) into Turkish.
Translated CFQ-Rs included the ones for children aged between 6 and 11, 12 and 13, 14 and above and for parents of children aged between 7 and 13. The process started with translation of CFQ-Rs into Turkish by two independent native Turkish speakers. Then each question and answer in the two translations was compared on a telephone conference with the author who has developed the form. Turkish translation is finalized taking the best of the two translations. Finally back-translation of the consensus Turkish CFQ-R is performed and reviewed on a telephone conference again. Finally to make sure it is applicable to Turkish children and parents, ten CF patients from each age group and ten parents were interviewed with one of the researchers. Last changes were made after this interview.
At the end of the questionnaire, all four CFQ-Rs have been translated and ready for use in Turkish children with CF. Validation and reliability study is still going on.
Translation of CFQ-Rs into Turkish is expected to aid medical doctors and other personnel in the care of children with CF.

315 Are the patient-reported outcomes (PROs) of value in annual review (AR) of adult patients with CF?

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Little attention has been paid to PROs such as HRQoL in the management of adult CF.
Aims: to compare measurement properties of a CF-specific measure, CFQoL, and a generic HRQoL instrument, UKSIP, in an attempt to establish their relative value as part of AR.
Methods: Patients with CF were recruited into the study from inpatient and outpatient clinics of a specialist Adult Centre for CF and by post for those who were not scheduled to attend the clinic during the study period. Patients were asked to complete CFQoL and UKSIP at baseline (Test 1) and again at 7–10 day intervals (Test 2).
Results: 118 were entered into the study of whom 70 took part (mean age 26, median 23, range 17–61 years; 45 male). 39 (56%) were unemployed, 82% of whom attributing it to their CF. The areas of HRQoL most impaired by CF were employment (UKSIP) and had concerns regarding their future (CFQoL). Psychosocial activities, particularly emotional behaviour, were more impaired than physical ($p < 0.05$). UKSIP and CFQoL showed high reliability in the CF population; Cronbach's alpha internal consistency coefficient (0.81–0.93) and test-retest correlations ($rs = 0.57$–0.94, $p < 0.005$). HRQoL scores reflected patient-rated overall health (very good to very poor-$p < 0.05$), showing discriminant validity. Females reported poorer HRQoL in all UKSIP categories ($p < 0.01$). UKSIP scores correlated with age ($p < 0.01$). CFQoL showed differences between severe and mild disease and BMI ($p < 0.01$).
Conclusion: The findings of the study indicate that CF has a large impact on HRQoL, and its assessment as part of CF annual reviews is recommended to provide holistic patient care and may positively influence management of CF.

318 The role of parental supervision on medical adherence in adolescents with Cystic Fibrosis

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Background: Previous research suggests that both parental supervision and adherence decrease in adolescence, as the drive for independence and autonomy emerge naturally during this period. However, no studies have examined this important relationship in adolescents with cystic fibrosis (CF).
Participants and Methods: The current study included 103 adolescents with CF (M age = 13.4, 47% female, M FEV1% predicted = 82.1%, 94% Caucasian). Activity patterns (medical and non-medical) were measured using the Daily Phone Diary and adherence to nebulized medications was measured electronically.
Results: Controlling for sex and FEV1, significant differences were found for time spent in medical care activities [Hotelling's $T = 0.11$, $F(2,98) = 5.3$, $p < 0.01$], with younger adolescents spending more time with parents in medical activities than older adolescents ($M = 32.0$ vs. $M = 17.6$ minutes). Results also indicated that time spent with maternal but not paternal caregivers ($\beta = 0.38$ vs. $\beta = -0.06$) was the most significant predictor of better adherence to nebulized medications.
Conclusions: Results from this study indicated that parental supervision of CF medical treatments declines with age, with maternal supervision being a significant predictor of adherence to nebulized medications. Overall these data suggest that parents and adolescents are likely to benefit from anticipatory guidance about how to transition responsibility, as well as how to facilitate independence while also rewarding good disease management.
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